Anomalous Origin of the Left Coronary Artery (AAOLCA) from the Right Coronary Sinus in a Young Male with Duchenne Muscular Dystrophy (DMD)

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Case Description

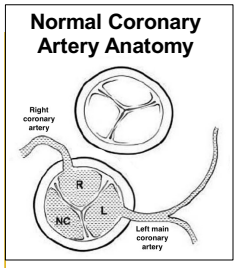
A currently 9-year-old patient with **Duchenne Muscular Dystrophy (DMD)** with an incidental finding of **anomalous aortic origin of the left coronary artery (AAOLCA) from the right coronary sinus** with an inter-arterial course.

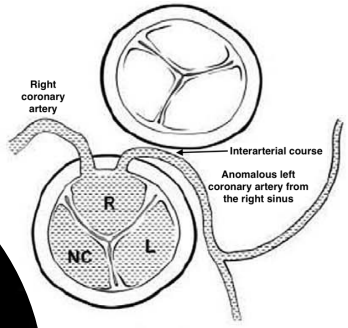
He was diagnosed with DMD in infancy due to a family history (older brother). The AAOLCA was initially found by routine transthoracic echocardiography. CT angiography confirmed the diagnosis and inter-arterial course of the anomalous coronary artery. He remains ambulatory and not reliant on positive pressure ventilation. He has had no significant systolic or diastolic cardiac dysfunction. The patient has had no concerning cardiac symptoms, including no chest pain, dyspnea, palpitations, easy fatigability, pre-syncope or syncope with or without exertion. He is very active and reportedly keeps up with his peers. His electrocardiogram shows sinus rhythm with a minor right ventricular conduction delay and no significant ST segment abnormalities.

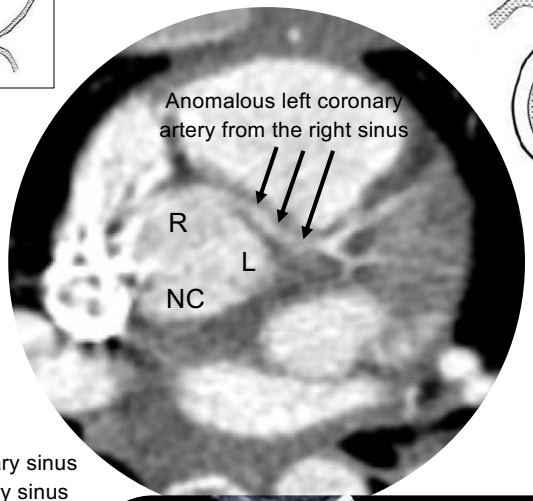
Though functional cardiac involvement is well described in dystrophinopathies, there are no documented associations between human dystrophinopathies and anomalous coronary artery origins (or other congenital heart disease).

DMD Pathophysiology

DMD patients undergo **progressive muscle degeneration** and weakness largely secondary to myocyte conversion to fibrofatty and connective tissue as well as mitochondrial dysfunction. **Physical activity decreases steadily with loss of ambulation**. Ultimate demise is most often due to progressive dilated cardiomyopathy and fatal arrhythmia burden. **DMD patients are at increased risk of cardiopulmonary compromise during anesthesia** due to a combination of cardiomyopathy, weak airway muscles and risk for rhabdomyolysis, hyperkalemia, and hyperthermia.

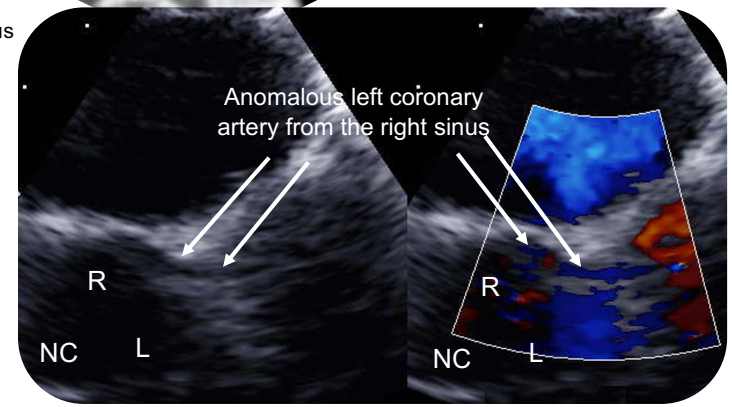






CT Angiogram

R – right coronary sinus
L – left coronary sinus
NC – non-coronary sinus



Echocardiogram

AAOLCA Pathophysiology

There is an approximate **6.3% incidence of sudden cardiac death (SCD) in AAOLCA**, the mechanism for which is not clearly defined but is most likely ischemic in nature. **SCD frequently occurs during or shortly after exercise**. It is thought that there is insufficient coronary blood flow to meet myocardial oxygen demand, which leads to ischemia and subsequent fatal ventricular tachyarrhythmias. High risk features include narrow coronary ostium, "slit-like" coronary ostium, acute coronary artery angulation, inter-arterial segment, and intramural course (which are thought to be compressed during systolic expansion of the great vessels during exercise). Recommended management for patients with AAOLCA is surgical correction, which has favorable peri-operative outcomes and low operative mortality. **Reported risk of post-operative inducible ischemia, however, is 37% and of SCD is 1.5%**.

Discussion

In a patient with DMD who is not expected to participate in competitive athletics but who is still ambulatory and active, management of his otherwise potentially high risk inter-arterial AAOLCA is not straightforward. **His presumed mortality is likely far less than a typical patient, given his lower expected level of physical exertion and decreased myocardial demand. Additionally, he is at higher peri-operative risk than a non-DMD patient**. It is unclear if the eventual cardiovascular changes associated with his dystrophinopathy will increase his risk for ischemic incidences related to his AAOLCA (or, for that matter, from its repair). There is evidence that coronary dilation potential is diminished in patients with dilated cardiomyopathy, which may be protective in the setting of a proximal stenosis. **Unfortunately, the longer we wait to decide if he would benefit from surgical repair (using risk stratification techniques), the higher his surgical risk becomes due to cardiopulmonary decline**.